Restricted mandibular movement associated with masseter muscle ossification in a patient with Fibrodysplasia ossificans progressiva
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Abstract
Fibrodysplasia ossificans progressiva is an extremely rare genetic disease which is transferred as an autosomal dominant gene and characterized by soft tissue ossification. This paper report an uncommon case of fibrodysplasia ossificans progressiva with ossification of the masseter muscle associated with osteomyelitis of the mandible.

Key Words: Fibrodysplasia Ossificans Progressiva; Heterotopic Ossification; Dental Complications

Received on: 22/11/2010    Accepted on: 12/01/2011

Introduction
Fibrodysplasia ossificans progressiva (FOP) is an extremely rare genetic disease characterized by progressive ossification of muscular system and connective tissue.(1, 2) FOP affects approximately 1 of 2 million people.(2) This disease usually begins in the first decade of life. In many cases, injuries can cause joints to become immobile in place and initiate development of a painful inflammatory mass. Although TMJ is one of the last joints affected by FOP, TMJ involvement reported in approximately 80% of patients by the age of 18.(3) Even non-complicated dental surgeries could develop ossification of masticatory muscles and consequently limited jaw mobility.(4) The aim of this article is to illustrate clinical and radiological features of FOP and dental complications in a rare case of FOP with an important degree of mandibular movement restriction.

Case Report
A 26 year old man was referred by his family physician to the orthopedic surgery clinic for evaluation. The patient reported tender and painful mass on the scapula without any history of local trauma since the age of three years. The patient was born by caesarean operation after an uneventful full term gestation. The parents and two younger brothers of the patient didn’t show any obvious skeletal malformation and they were reported as normal. At that time, the patient was referred to general surgeon by his pediatric physician to remove the mass which was diagnosed as soft tissue tumor. After surgical removal of the lesion, the patient experienced stiffness and restricted movement of the neck. The patient had another surgery on the same region two months later because of increased limitation of neck extension. The patient stated that he has been suffered from stiffness and restricted movement of spine, knees, shoulders, hip and neck for the past 20 years. Physical examination revealed normal cardiovascular system without any complication. All Laboratory data including hemogram, erythrocyte sedimentation rate, serum calcium and phosphorus, alkaline phosphatase, creatinine phosphokinase, routine urinalysis, urine calcium and phosphorus, and creatinine clearance were within normal limits. Lateral neck radiograph showed ossification of trapezius muscle as well as cervical spine fusion (Fig 1). Hand radiograph of the patient showed shortening of the first metacarpal of the fifth finger (Fig 2). Conventional radiograph of the lower limb showed ossification of the quadriceps muscles (Fig 3).

Fig. 1
Fig. 2
Fig. 3
Magnetic resonance and imaging (MRI) of neck showed no evidence of lymphadenopathy in parapharyngeal space. MRI of the spine showed multiple ossifications of varying sizes along the paraspinal muscles. Based on clinical and para-clinical findings, diagnosis of FOP was made.

The patient was referred to the Department of Oral and Maxillofacial Surgery due to a painful swelling present over the left cheek region and restriction of his mouth opening. Temporomandibular joints were not tender on palpation. Initial examination revealed a maximum opening of 18 mm with deviation of the mandible to the left side on opening. Oral examination of the patient showed a swelling at the buccal space of the left mandible and multiple dental caries. Unfortunately, the patient did not get treated for dental caries and mandibular infection because of the fear that dental procedures may worsen the disease. The patient underwent short period of antibiotic therapy by his family physician for mandibular infection. Radiographic examinations revealed irregular bony appositions over the mandibular ramus of the left side, extending from the lateral surface of the zygomatic bone toward the angle of mandible, with destruction of cortical and spongy bone of the left mandible. Owing to the patient’s physical condition which had prevented him from undergoing mandatory dental care and short period of antibiotic therapy, the uncontrolled infection was developed into mandibular osteomyelitis. Afterward, ossification of the left masseter muscle occurred which was most likely responsible for the restricted mandibular movement. There was no report of maxillofacial trauma in the medical history of the patient. These findings have led to diagnosis of the ossification of the masseter muscle due to mandibular osteomyelitis whereas infection is a risk factor for ossification of muscles.

Discussion

Guy Patin first described FOP in 1648 in a young woman who “turned to wood”. The autosomal dominant inheritance of FOP was first described by Sympson in a case report of a seven year old boy with classic features of FOP.(5) Ossification of muscular system usually occurs from birth up to the age of 16 (mean age 4.6 years) following spontaneous or trauma-induced “flare-ups”.(2, 5) Heterotopic ossification usually begins in the cervical paraspinal muscles and later spreads from axial to appendicular, cranial to caudal and proximal to distal regions.(4, 5)

Malformation of the great toes and progressive heterotopic ossification of muscles, ligaments and tendons are the main clinical manifestations of FOP.(5) In the present case abnormality of the great toes, with shortening of the first metatarsal and proximal phalanx, was observed. In addition, painful swelling of the scapula without any history of trauma was reported since the age of three years.

Conductive hearing impairment is a common manifestation of this disease and it probably occurs due to the fusion of the ossicles of the ear. Cardiopulmonary complications are common in the older patients.(4, 5) There was not any report of cardiopulmonary problems as well as hearing impairment in the present patient. Despite the extensive knowledge of metabolic bone disease, there is no standard treatment protocol for FOP. Treatment of FOP is multifactorial and based on injury prevention and clinical therapy. Effective treatments for FOP and other heterotopic ossification conditions depend on future discoveries that inhibit ACVR1 (also known as activin-like kinase) signaling.(2, 3) Inaccurate diagnosis of this disease can leads to permanent injury and changes the natural history of the disease, as seen in the present case.(6)

Laboratory tests may reveal a discrete increase of erythrocyte sedimentation rate during the “flare-ups”. Additional Imaging such as MR imaging, bone scanning and CT scans are necessary to achieve accurate diagnosis.(5) Previous reports identify trauma as one of the most common causes of the heterotopic ossification of muscles and disease reactivation.(5) However, maxillofacial trauma was not reported in our patient. In the present case, mandibular infection may have initiated ossification of the masseter muscle.

Surgical treatment is risky in FOP patients and causes recurrence and exacerbation of disease within few months. In case of mandatory surgical operations, anesthesiologist should be evaluate special considerations for the patient with FOP such as neck immobility, limited jaw opening.
superficial intravenous access and general anesthesia with fiberoptic nasotracheal intubation. Restricted mandibular movement is the major manifestation of FOP in the maxillofacial region and occurred in approximately 80% of the patients. (2, 5, 7)

In order to avoid long-term dental complications, preventive oral health care and regular checkups are crucial for the patients with FOP especially during primary dentition period. Frequent brushing and flossing and use of high dose fluoride toothpaste are strongly recommended, along with use of chlorhexidine rinses and fluoride therapy to help prevent dental decay and periodontal disease. In patients who are unable to open their jaw adequately, special toothbrushes as well as antibacterial and fluoride rinses may be useful. (3, 8)

Intramuscular injections including vaccines and mandibular block anesthesia should be avoided. Intra ligamentary and infiltration anesthesia are not contra-indicated in patients with FOP. (3) Dental practitioners should avoid overstretching of the mandible during any dental procedures. (8)

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Source of Support: Nil, Conflict of Interest: None Declared